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Case Report

A Case Report of Non-specific Symptoms of Takayasu's Arteritis in a 17-year-old Female Patient

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ABSTRACT

Takayasu's arteritis (TA) is a major chronic vasculitis disorder that its etiology is unknown. Patients are mostly Asian women who often show nonspecific symptoms such as fever, myalgia, arthralgia, weight loss, and anemia. The report relates to a 17-year-old girl suffering from complaints, permanent and uncontinental pain starting from a month earlier, with loss of appetite and weight loss (4 kg), and night sweats. She had no diarrhea or gastrointestinal symptoms but had a pain in the shoulder and chest area since the last 5 months. She got better after seeing a physician and receiving supplements. She had a history of pain in the ear from the last five months leading to otitis, and was treated as a case of brucellosis with a score of 1.20 in the Coombs Wright test. According to the physical examination findings, the patient's left, radial, ulnar, and proximal pulses, and blood pressure were unexplained, and in the supraclavicular region of the left and the umbilical region, bruit was heard and the shape of the left nail was changed. Laboratory tests and imaging were performed for the patient, and after angiography, the left subclavian artery stenosis was detected. Given the age and sex of the patient and the results obtained, she was diagnosed with Takayasu's arteritis.

Keywords: Takayasu's arteritis (TA), Large-vessel vasculitis, Sedimentation rate (ESR)

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Introduction

Takayasu's arteritis (TA) is a panarteritis which its etiology is unknown. It usually affects aorta, major pulmonary arteries and its branches (1). TA is called pulseless disease; it is an atypical origin of symptomatic peripheral vascular disease (2). Based on the history of TA, early accounts of patients were found out in 1856, 1872, and 1908 but an overall discussion of pulseless disease was not offered until 1984 (3). It was indicated as a much more predominant disease than previously thought. It was more extensive among young Japanese women, however, it is now an international disease affecting all age groups, and has various patterns of clinical and angiographic presentation (2). Protean clinical manifestations at times avert us from a certain diagnosis for a long time. It is due to the fact that inflammatory condition circumspectly leads to either stenosis occlusion and/or dilatation of the involved arteries. So, increasing awareness of the diseases is of great significance (4, 5).

This report describes a case of Takayasu's arteritis with cut off left subclavian artery and right renal artery stenosis (99%) in a 17-year-old girl who visited a physician with back and chest pain, fever and by 1:20 titer in Coombs Wright test as Brucellosis had been treated.

Case Report

A 17-year-old girl referred to a physician with fever, back and chest pain. She reported that the pain had been with her since six months earlier. It was a steady and constant pain since one month before which was not related to eating habits and not a positional pain. She had anorexia and lost weight from 59 kg to 55 kg. She also suffered from night sweating. She did not have diarrhea or any other GI presentations but in the previous week, she once had nausea and vomiting. She also mentioned Hx of neck pain, shoulder pain that had been radiated to intrascapular space, and chest pain since five months ago. In the beginning, the nature of pain was on/off and took approximately five minutes. She had taken calcium supplement after consulting with a physician, and she got better.

However, three months earlier, the pain returned and she visited another physician. She had been eating rural diary and had been treated by 1:20 titer in Coombs Wright test as a Brucellosis. She hinted at her left ear tinnitus since five months earlier due to otitis. She also had an adenectomy surgery when she was nine.

In the process of examination, the left radial, ulnar, and brachial pulses and BP were not detected. The blood pressure in the right arm was 145/100. She had a bruit on the left supraclavicular region and umbilical zone. The shape of index finger nail on the left hand had been changed. The laboratory parameters included a white blood count of 9400 per µl, hemoglobin of 9.7 g/dl, and platelet count of 527/00 per µl. The erythrocyte sedimentation rate (ESR) was 120 mm/h. CRP was 61. The results of Anti-CCP, Anti-ds-DNA, and Wright agglutination tests were all negative. In the renal function tests, urea was 15 and Cr was 1.1. Echocardiography was performed and the results were normal. The Compound Tomography (CT) of abdomen and pelvic was unremarkable. Doppler sonography of kidney and carotids arteries were normal. The results of angiography showed cut off left subclavian artery (Figure 1) and 99% stenosis of right renal artery (Figure 2). Although the aorta seemed normal, the nature of narrowing of involved arteries along with age and gender of the patient appeared to be consistent with TA.

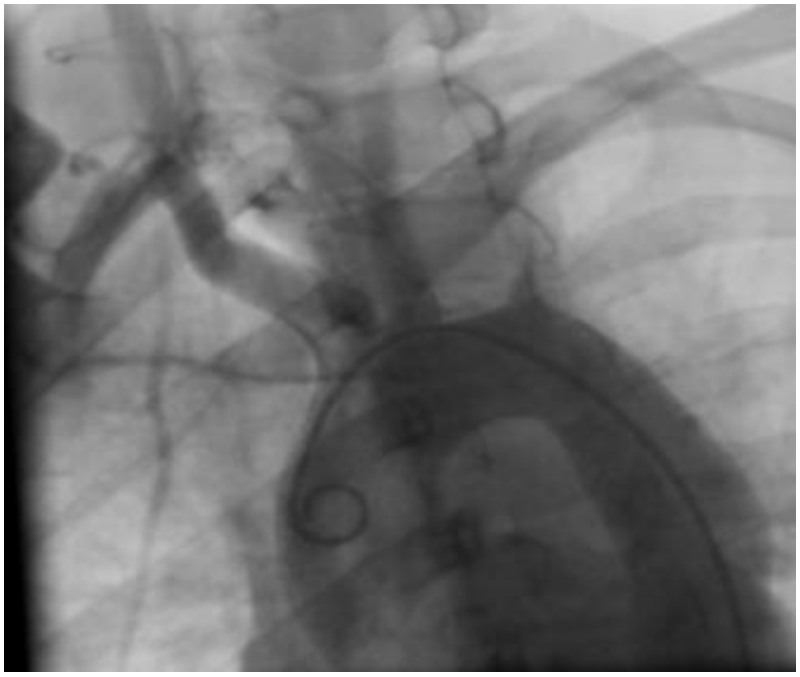

Figure 1. Angiography of subclavian artery that demonstrates cut off left subclavian artery.

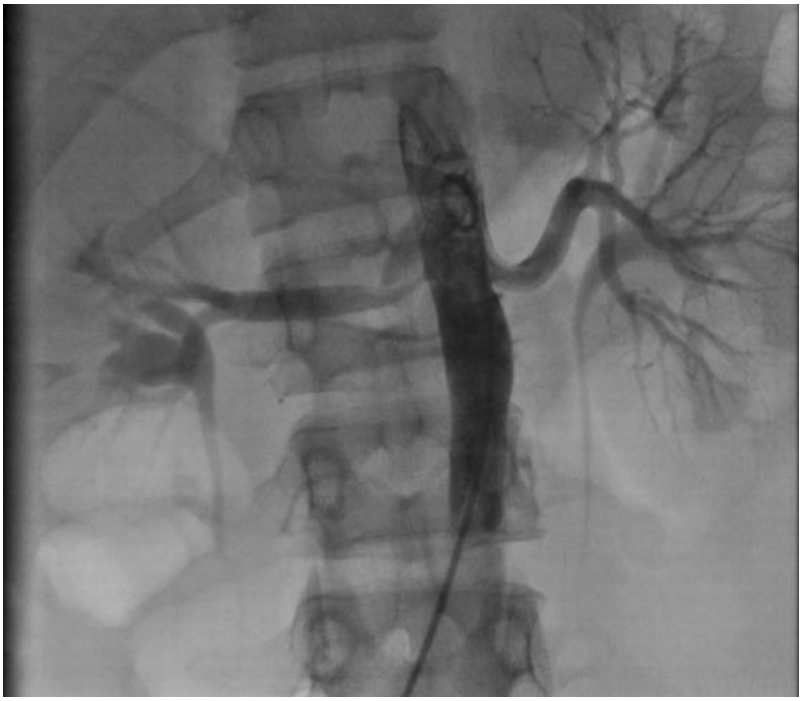

Figure 2. Angiography of renal artery that shows 99% stenosis of the right renal artery.

Discussion

Takayasu's arteritis (TA) is an uncommon disease and the epidemiologic findings differ based on the geographical state. It was previously considered to affect young women in Far East Asia, but it is now evident that it is able to affect all ethical groups both men and women at different ages all over the world (6). Japan has the highest generality described in TA (40 cases per million) (7).

Genetic studies proved that HLA-B52 and less frequently B67 in Japan are the most significant HLA alleles related to TA in various ethnicities (8). We have considerably improved our information of TA in the last decade, but the exact pathogenic sequence is not clarified yet. The improved knowledge of the immunological

mechanisms, which are responsible for vascular injury in TA has brought about trials of anti-TNFX agent with motivating results (9). Grayson et al. proved that TA has the highest rate of new, severe, and various vasculitises among six different vasculitises (ischemic, vascular) incidence (44%) (10).

Systematic symptoms of fatigue, loss of weight, and low grade fever are frequent in the first phase and vascular symptoms are not widespread. Then, patients look chronically sick. A number of vascular symptoms appear as the result of narrowing, occlusion, and dilatation of the affected arteries (2). There were six principles in the traditional format grouping: Onset age < 40 years, claudication of an extremity, decreased brachial artery pulse, > 10 mmHg difference in systolic blood pressure between arms, a bruit aorta, its primary branches, or large arteries in the proximal upper or lower extremities. When three or more principles were reported, there was a sensitivity of 90.5% and a specificity of 97.8% (3), that our case had all the above-mentioned principles.

The basis of clinical diagnosis of TA were clinical and angiographic data (1).

The awareness of radiographic, computed tomographic, magnetic resonance imaging (MRI), and angiographic characteristics of TA may pave the way for an earlier diagnosis and treatment (2).

Although the most sensitive test for early vessel inflamation is 18-FDG-PET. If 18-FDG-PET shows activity, it is considered as a hint to active or relapse the disease that lessens after immunosuppression (7). Moreover, CT and in recentup (11).

Considering the prognosis of TA in one of the biggest series that has a long follow-up from Mayo Clinic in the US, general indurance has improved significantly in comparison to the earlier series (97% at the age of 10 and 86% at the age of 15) (12). It must be kept in mind that the leading goal of the treatment of TA is inspiring cancellarion and control of acute vasculitis by pharmacologic agent. Glucocorticoids are the main drugs for treatment of TA, and similar to other vasculitis (6), different cytotoxic drugs such as methotrexate and cyclophosphamide have been used with variable success (1). Contemporary unrestricted data of leflunomide, TNFx-antagonists and tocilizumab in refracting TA look favorable (13). Surgical approaches and interventional radiological way may be applied for aomplications of stenosis, occlusions or aneurysms in TA (6).

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